

Amendments to the Claims:

1. (Currently amended) ~~A transgenic Transgenic~~ non-human animal;
~~preferably a knock-in mouse, having~~ comprising a missense mutation in the $\alpha 4$ - or $\beta 2$ -
subunit of the neuronal nicotinic acetylcholine receptor (nAChr).

2. (Currently amended) The animal ~~Animal~~ of claim 1, ~~having the wherein~~
said the missense mutation is V287L or V287M in the gene ~~for~~ encoding the said $\beta 2$ -
subunit of said nAChr receptor.

3. (Currently amended) The animal ~~Animal~~ of claim 1, ~~having the wherein~~
said the missense mutation is selected from the group consisting of 259-260ins, S252L,
766ins3 ~~or~~ and T265I and said mutation is in said $\alpha 4$ -subunit of the said nAChr receptor.

4. (Currently amended) The animal ~~Animal of any one of claims claim 1 to 3~~
containing, wherein said animal is homozygous for said the missense mutation
homozygously.

5. (Currently amended) ~~The animal~~ Animal of any one of claims claim 1 to 3 containing, wherein said animal is heterozygous for said the missense mutation heterozygously.

6. (Currently amended) A targeting ~~Targeting~~ vector containing comprising the following components operatively linked: the a genomic and/or cDNA nucleic acid sequence for encoding a subunit of the, preferably a human or murine, nicotinic acetylcholine receptor (nAChR) having a missense mutation in the $\alpha 4$ - or $\beta 2$ -subunit, or a part of said subunit, wherein said part has comprises at least the said missense mutation in the $\alpha 4$ - or $\beta 2$ -subunit operably linked to a selectable marker gene, and ~~optionally 2 recognition sequences for a recombinase, which flank the marker~~ gene.

7. (Currently amended) The targeting ~~Targeting~~ vector of claim 6, wherein the selectable marker is an antibiotic resistance gene.

8. (Currently amended) The targeting ~~Targeting~~ vector of claim 6 or 7, wherein said vector further comprises ~~recognition sequences~~ two 2 recognition sequences for a recombinase that, which flank the marker gene ~~are each loxP~~.

9. (Currently amended) ~~The targeting~~ Targeting vector of ~~claim one or more of claims 6 to 8~~, wherein the β 2-subunit ~~has the~~ comprises a ~~missense mutation V287L or V287M~~ missense mutation.

10. (Currently amended) ~~The targeting~~ Targeting vector of ~~one or more of claims claim 6 to 8~~, wherein the α 4-subunit has the a missense mutation selected from the group consisting of 259-260ins, S252L, 766ins3 or T265I.

11. (Currently amended) ~~A stem~~ Stem cell, ~~preferably murine embryonic stem cell, containing~~ comprising a vector of ~~claim one or more of claims 6 to 10~~.

12. (Currently amended) ~~A screening~~ Screening method for the identification of compounds for the treatment of the human epilepsy syndrome, ~~particularly familiar nocturnal frontal lobe epilepsy (ADNFLE)~~, comprising the following steps:

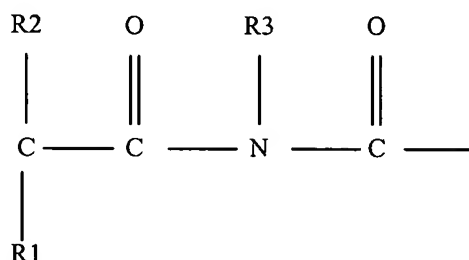
a) providing an animal ~~of any one of claims claim 1 to 5~~ and providing a test ~~compound~~ compounds,

b) administering said ~~administration of the test compound~~ compounds to the said animal,

c) ~~selecting selection of a test compound resulting in that alleviation or elimination of the~~ alleviates or eliminates symptoms of the an epilepsy syndrome in the said animal, and

d) ~~optionally repeating the steps a) to e) with a suitably modified form of the test compound chosen in e).~~

13. (Currently amended) ~~The screening~~ Screening method of claim 12, wherein the said test compound is ~~compounds~~ are selected from the group consisting of ~~following groups~~ barbiturates, oxazolindiones, ~~and succinimides,~~ derivatives of benzodiazepines, sultiam, Carbamazepin, valproic acid and compounds ~~further groups having the following grouping as a group comprising as a common structural element~~ formula I:



wherein R^1 and R^2 are alkyl or aryl residues and R^3 is H or an alkyl residue,
~~or derivatives of benzodiazepines, sultiam, Carbamazepin and valproic acid.~~

14. (Currently amended) A compound ~~Compound~~ for the treatment of the a human epilepsy syndrome, preferably of ADNFLE, which has been identified by according to the method of claim 12 or 13.

15. (Currently amended) A pharmaceutical ~~Pharmaceutical~~ composition having comprising a therapeutically effective dose of one or more a compound identified according to the method of claim 12 in compounds of one or more of claims 12 to 14 and a pharmaceutically acceptable carrier.

16. (Currently amended) Use of the composition of claim 15 for the treatment of the A method for treating a human epilepsy syndrome, preferably ADNFLE by administering to a patient a composition identified according to the method of claim 15.

17. (New) The transgenic non-human animal of claim 1, wherein said animal is a knock-in mouse.

18. (New) The targeting vector of claim 6, wherein said nucleic acid is a genomic and/or cDNA nucleic acid sequence.

19. (New) The screening method of claim 12, where in said human epilepsy syndrome is familial autosomal dominant nocturnal frontal lobe epilepsy (ADNFLE).

20. (New) The screening method of claim 12, wherein said method further comprises:

d) repeating steps a) to c) with a modified form of the test compound chosen in c).